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Case report

Multiple intramuscular myxomas associated with polyostotic fibrous dysplasia

Koichi Fujii*, Masaaki Inoue, Yutaka Araki, Osamu Ishida

Department of Radiology, Kinki University School of Medicine, Osaka, Japan

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1. Introduction

Intramuscular myxoma is one of the rare but important associations of skeletal fibrous dysplasia. The association has been seen most frequently with polyostotic fibrous dysplasia, and occurs years after fibrous dysplasia has presented in most cases.

The clinical behavior and pathological appearance of soft-tissue myxoma are sometimes mistaken for myxoid liposarcoma.

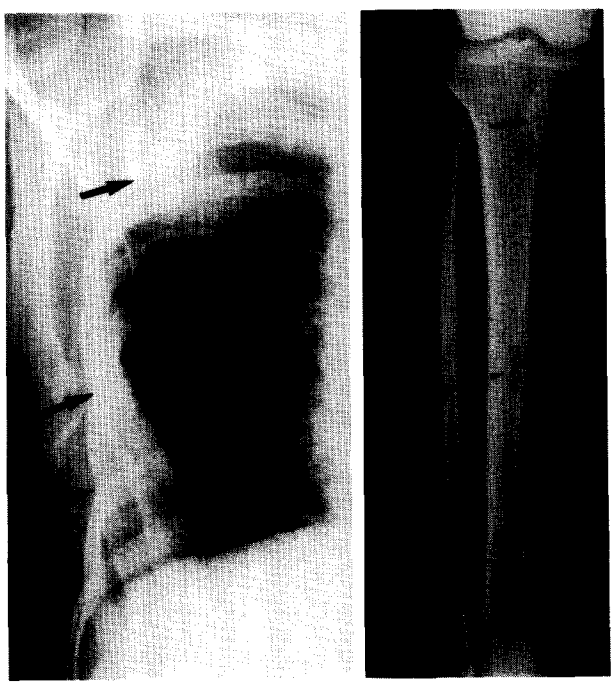
A case of multiple intramuscular myxomas of the bilateral hips and right thigh, associated with polyostotic fibrous dysplasia of the right tibia and right ribs is presented.

2. Case report

A 53-year-old woman presented with a painful mass in the right hip in May 1990 and a painless mass in the right thigh in August 1990. She had a history of skeletal abnormalities in the right tibia and right ribs since 1987.

Physical examination revealed a 8 × 8 cm painful mass in the right hip, a 2 × 2 cm painless mass in the right thigh and a 3 × 3 cm painless mass in the left hip. The patient was unaware of the mass of the left hip. There were no skin lesions such as café au lait spots. Laboratory data including endocrinological findings were entirely normal.

A radiograph of the right rib showed multiple expansive ground glass lesions (Fig. 1a). The right tibia presented a radiolucent lesion with marginal sclerosis in the proximal diaphysis and a radiolucency without marginal sclerosis in the metaphysis (Fig. 1b). A



a b

Fig. 1. (a) Radiograph of the right rib showing multiple expansive ground glass lesions (black arrows) of the 2nd, 6th and 7th rib. (b) Radiograph of the right tibia showing a radiolucent lesion (black arrow) with marginal sclerosis in the proximal diaphysis and a radiolucency without marginal sclerosis (black arrowhead) in the metaphysis.

* Corresponding author, Tel.: +81 723 660221 (Ext. 3132); Fax: +81 723 682388.

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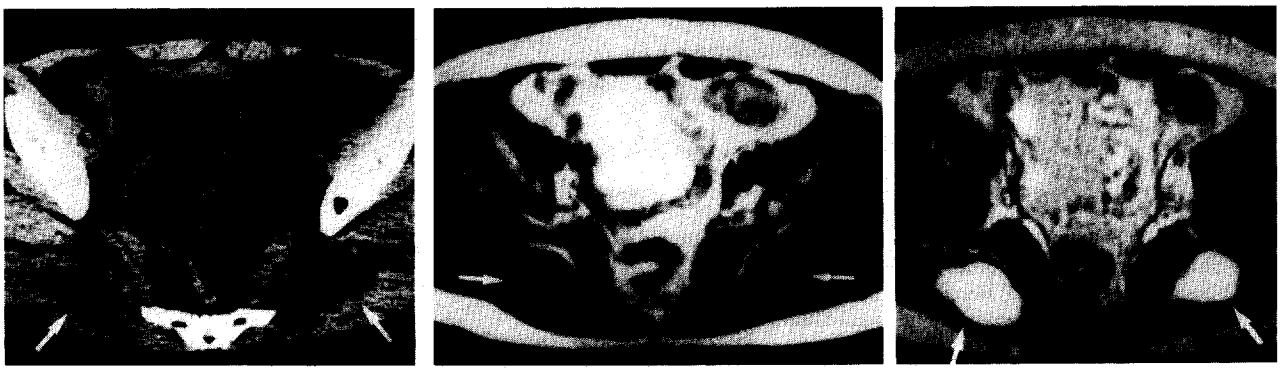

a b c

Fig. 2. (a) CT scan reveals oval water density lesions (white arrows) in the bilateral hips. (b) MR axial T1-weighted and (c) T2-weighted images demonstrate the lesions (white arrows) with water signal intensity in the bilateral hips.

technetium bone scan showed increased uptake coincident with radiographical lesions of the right rib and right tibia. No extraskeletal uptake was seen.

Computed tomography (CT) revealed oval water density lesions in the bilateral hips (Fig. 2a). Magnetic resonance (MR) imaging of the hip demonstrated the lesions with water intensity on both T1-weighted and T2-weighted images (Fig. 2b,c). The lesion of the right thigh also showed the same findings on CT and MR.

Incisional biopsy of the proximal lesion of the right tibia was performed. Microscopically, the lesion showed irregular woven bone trabeculae surrounded by abundant fibrous matrix. The lesion of the right hip was excised at surgery. Histologically, the mass was hypocellular, consisting of scattered stellate cells lying in large amounts of mucoid matrix with poor vascularity. The diagnosis of fibrous dysplasia of bone and soft-tissue myxoma was confirmed pathologically.

3. Discussion

Intramuscular myxoma is a rare benign tumor arising from fibroblasts that produce excessive amounts of mucopolysaccharide. The myxoma is usually painless, presenting as a mass between the fourth and seventh decades, and most commonly occuring in the thigh and shoulder [1]. On CT and MR, intramuscular myxoma shows a well-defined, homogeneous, oval mass having an attenuation and a signal intensity equal to or close to water [1–6].

The association of intramuscular myxoma with fibrous dysplasia was first described in 1926 [7] and the importance of recognizing this association was emphasized by Mazabraud et al. in 1967 [8]. Wirth et al. [9] in 1971 called attention to this association as a largely overlooked manifestation of fibrous dysplasia.

A review of the literature reveals that only 25 cases of the association have been reported. Of the 26 cases including our case, 11 were men and 15 were women. The diagnosis of fibrous dysplasia ranged from 1 to 57 (mean 22) years, 87% within the first three decades of life. The presentation of intramuscular myxoma ranged from 17 to 82 (mean 46) years, 68% older than 40 years. In most cases, intramuscular myxoma tended to occur many years later. Five cases of this association were with in a monostotic form of fibrous dysplasia and 21 were with in polyostotic forms. Fifteen cases of fibrous dysplasia involved the unilateral side of the body and 11 involved the bilateral side. In 13 of the 15 cases with unilateral fibrous dysplasia, myxomas occurred in the ipsilateral side of the adjacent soft tissue. The myxomas were multiple in 20 of 26 cases and showed a predilection for the right side of the body in 15 cases [3,5,6–13].

In clinical, radiological and pathological appearance, intramuscular myxomas may be difficult to distinguish from myxoid containing tumors, especially myxoid liposarcomas [1,2,4,10,14]. In some of the cases, there may be also scattered macrophages with small intracellular droplets of lipid material [13]. Therefore, intramuscular myxomas can be confused with myxoid liposarcomas. Myxomas, however, have poor vascularity compared with myxoid liposarcomas having abundant vascularity.

As regards the etiology of intramuscular myxoma, Wirth et al. considered soft-tissue myxomas an extraskeletal manifestation of fibrous dysplasia and presumed a basic inborn error of tissue metabolism as the cause [3,5,10,12]. However, the hypothesis alone cannot always explain why few patients with fibrous dysplasia develop myxomas. The mechanism of this association needs further investigation.

Although fibrous dysplasia seems to be unrelated to soft-tissue myxoma, radiologists need to be aware of this rare but important association. If the patients with fibrous dysplasia develop soft-tissue masses, intramuscular myxomas should be firstly suspected. In

contrast, if the patients have soft-tissue myxomas, skeletal surveys should be necessarily done considering fibrous dysplasia.

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